Differences in social motivation in children with Smith-Magenis Syndrome and Down Syndrome

Wilde, Lucy; Oliver, Chris; Mitchell, Anna

DOI:
10.1007/s10803-016-2743-3

Document Version
Peer reviewed version

Citation for published version (Harvard):
Wilde, L, Oliver, C & Mitchell, A 2016, 'Differences in social motivation in children with Smith-Magenis Syndrome and Down Syndrome', *Journal of Autism and Developmental Disorders*, vol. 46, no. 6, pp. 2148–2159. https://doi.org/10.1007/s10803-016-2743-3

Differences in social motivation in children with Smith-Magenis syndrome and Down syndrome

Lucy Wilde, Anna Mitchell and Chris Oliver

Cerebra Centre for Neurodevelopmental Disorders,

School of Psychology,

University of Birmingham

Please use this reference when citing this work:

Wilde, L., Mitchell, A., & Oliver, C. (2016). Differences in social motivation in children with Smith-Magenis syndrome and Sown syndrome. *Journal of Autism and Developmental Disorders*.

The Cerebra Centre for Neurodevelopmental Disorders,

School of Psychology, University of Birmingham, Edgbaston, Birmingham, B15 2TT

Website: www.cndd.Bham.ac.uk **E-mail**: cndd-enquiries@contacts.bham.ac.uk

Abstract

Social excesses, characterised by heightened social motivation, are important for describing social functioning. Smith-Magenis syndrome (SMS) is a potential exemplar of a disorder where heightened social motivation is associated with negative behavioural outcomes. In Down syndrome (DS) strong social motivation is described, but less commonly associated with behavioural problems. Children with SMS (n = 21) and DS (n=19) were observed during social situations, in which familiarity of adults present and level of attention available were manipulated. Motivation in SMS was characterised by comparatively frequent social initiations when adult attention was low, and stronger preference for familiar adults, compared to DS. Findings provide insight into the nature of social motivation in SMS and support an argument for nuanced consideration of motivation.

Keywords; social motivation, neurodevelopmental disorder, Smith-Magenis syndrome, Down syndrome

Research into social behaviour in neurodevelopmental disorders has focussed on difficulties associated with deficits in social cognition and a range of social impairments in autism spectrum disorder (ASD) (Cook & Oliver, 2011; Moss & Howlin, 2009). However, some neurodevelopmental disorders demonstrate social excesses which are also potentially associated with negative outcomes. Specifically, heightened levels of behaviours that function to initiate or maintain social interactions. These behaviours are described as indicative of levels of social motivation (Chevallier, Kohls, Troiani, Brodkin, & Schultz, 2012). Early descriptions of ASD identified variation of social motivation, with some children described as indifferent to social situations ('aloof') and others as having greater social motivation, seeking social contact but in an inappropriate manner ('active but odd') (Wing & Gould, 1979). Further evidence for a spectrum of social motivation, encompassing excessive motivation, is provided by descriptions of elevated sociability in other neurodevelopmental disorders.

In Williams syndrome, caregiver reports of 'hypersociability' (Doyle, Bellugi, Korenberg, & Graham, 2004) are supported by behavioural observations. Behaviour during the Parental Separation task from the Laboratory Temperament Assessment Battery (LabTab) (Goldsmith & Rothbart, 1993), for example, indicated that children with Williams syndrome showed increased sociability on their parent's return compared to chronological or developmental age matched typically developing children (Jones et al., 2000). In Angelman syndrome, observations of elevated positive affect during social situations, particularly where high levels of adult attention are available, also implicate heightened social motivation (Oliver, Demetriades, & Hall, 2002; Oliver et al., 2007). Interestingly it is suggested that this motivation may be directed preferentially towards children's mothers (compared to unfamiliar adults) to a greater extent in Angelman syndrome than in children without this syndrome (Mount, Oliver, Berg, & Horsler, 2011).

Variability in the strength and nature of social motivation in neurodevelopmental disorders has implications for theoretical models of the development of atypical social behaviour. Reduced social motivation has been implicated in the aetiology of ASD (Dawson, Meltzoff, Osterling, Rinaldi, & Brown, 1998; Grelotti, Gauthier, & Schultz, 2002), with some ascribing it causative status in the development of deficits in social cognition (Chevallier et al., 2012). However, the implications of increased motivation within such models remains poorly understood. It may have different manifestations across disorders and is likely to be more nuanced than a broad 'impaired to excessive' continuum. Therapeutic implications are also likely. In Williams syndrome, social disinhibition results in inappropriate physical initiations and disruptive fixation of attention and affection on particular people (Davies, Udwin, & Howlin, 1998). In children with Angelman syndrome frequent grabbing and hair-pulling may function to access attention (Oliver et al., 2007). Delineation of social motivation in neurodevelopmental disorders with heightened motivation may provide more detailed understanding of motivation and its consequences.

One such candidate disorder is Smith-Magenis syndrome (SMS). SMS is caused by a de novo deletion on chromosome 17p11.2 or mutation of the retinoic acid-induced 1 (RAI1) gene and has a prevalence of 1/ 25,000 births (Greenberg et al., 1996; Slager et al., 2003; Smith et al., 1986). Key features include intellectual disability (ID), sleep disturbance, health problems and behavioural disorder (including self-injury and aggression) (Greenberg et al., 1996; Smith, Dykens, & Greenberg, 1998). Several sources implicate heightened social motivation in SMS and highlight the potential clinical impact. Caregivers report that over 80% of individuals with SMS show 'attention seeking' or 'demand a lot of attention' (Dykens, Finucane, & Gayley, 1997; Dykens & Smith, 1998). High levels of 'attachment' to particular favourite people are also reported (Moss, Oliver, Arron, Burbidge, & Berg, 2009). Strong social preference is reflected in naturalistic behavioural observations, in which children with

SMS showed stronger preference for attending to and interacting with adults (over peers) than children with Down syndrome (DS) (Wilde, Silva, & Oliver, 2013). Anecdotally, individuals with SMS are also described as demanding an 'inordinate amount of' and having a 'sometimes insatiable' need for individualised attention from adults (and little interest in peers), with aggression resulting if adult attention is restricted (Haas-Givler, 1994). This association between reduced attention and challenging behaviour is supported by indirect functional analytic studies (Langthorne & McGill, 2012; Sloneem, Oliver, Udwin, & Woodcock, 2011) and one direct assessment in which challenging behaviours were preceded by low levels of adult social contact for three of four children studied (Taylor & Oliver, 2008).

Studies of social behaviour in SMS have been limited by a lack of experimental methodology and small sample sizes. In this study we examine nuanced dimensions of motivation in SMS using experimental methods. Responses of children with SMS to systematic changes in the level of available attention (given reports of attention-seeking and negative response to reduced attention) during interactions with adults of differing familiarity (given reports of unusually strong preference for familiar adults) will be observed. These will be contrasted with those of children with DS to further delineate variability in social motivation in neurodevelopmental syndromes. Ability and language profiles in DS are comparable to those reported in SMS (Chen, Lupski, Greenberg, & Lewis, 1996; Greenberg et al., 1996; Martin, Klusek, Estigarribia, & Roberts, 2009; Melyn & White, 1973). Crucially, strong social motivation is also described. However, unlike in SMS this is less commonly associated with problematic behavioural outcomes. Instead, it is typically described as an asset (Hodapp, Ly, Fidler, & Ricci, 2001).

When matched on mental age, young children with DS attend more to caregivers' and strangers' faces than typically developing infants (Kasari, Mundy, Yirmiya, & Sigman, 1990) and show more social interaction behaviours than both typically developing children and children with ID (Mundy, Sigman, Kasari, & Yirmiya, 1988). Older children with DS are also more likely than others with ID to look to adults during problem-solving tasks (Kasari & Freeman, 2001). Furthermore, adults and children with DS rate social stimuli demonstrating happy emotions as more approachable than typically developing mental age matched controls, and at similar levels to 'hypersociable' individuals with Williams syndrome, although this difference was not evident for other emotions (Porter et al., 2007). However, compared to children with Williams syndrome, children with DS are more reserved towards strangers (Gosch & Pankau 1996b, as cited in Gosch & Pankau, 1997). Relative to other children with ID, they are also not rated as more socially competent (Griffith, Hastings, Nash, & Hill, 2010) and show significantly greater preference for being alone (63% compared to 28%) (Dykens & Kasari, 1997).

There is mixed evidence therefore as to whether social motivation is objectively elevated in DS (compared to mental age expectations) but, as with SMS, strong motivation is a key feature in the profile of behaviours described *within* the syndrome. Comparing SMS and DS therefore enables use of a 'same but different' approach to examining behaviour (Hodapp & Dykens, 2001), whereby two aetiologically different groups showing broadly similar behaviours are compared in order to highlight fine grained behavioural differences. Gross variation in social motivation has been established (e.g. within ASD, between neurodevelopmental syndromes). Here we extend this to compare two groups reportedly characterised by strong social motivation, to identify nuanced differences in the manifestation of motivation. Problematic attention seeking and very strong preference for specific (presumably familiar) people are described in SMS but not in DS. This suggests that

manipulating levels of available attention, and familiarity of social partners present during an interaction, may elicit greater changes in socially motivated behaviours in SMS than in DS. Identification of such differences may provide insight into why strong social motivation is reported to be particularly problematic in SMS.

The following hypotheses were therefore proposed;

The effect of decreased attention:

1) Given evidence of attention seeking behaviour in SMS, it was anticipated that in situations where attention is unavailable, children with SMS would seek to initiate interaction, reflecting increased social motivation in this situation. It was thus predicted that initiation of interaction when attention was unavailable (compared to available) would be greater in SMS than DS.

2) As social motivation in other syndromes (e.g. Angelman syndrome) is indexed by increased positive affect during social interactions, it was predicted positive affect when attention is available (compared to unavailable) would be greater in SMS than DS.

The effect of differing familiarity:

3) Given reports of preference for specific 'favourite' (therefore familiar) people in SMS, it was predicted that preference for initiation of interaction with mothers (compared to an unfamiliar adult) would be greater in SMS than DS, reflecting increased social motivation towards preferred adults.

4) As social motivation is expected to be greater towards mothers than unfamiliar adults, and positive affect is a potential index of motivation it was predicted that positive affect during interactions with mothers (compared to an unfamiliar adult) would be greater in SMS than DS.

Presence of differences between social motivation in children with SMS compared to DS in line with these hypotheses would be indicated by interactions between syndrome and experimental condition in analyses.

Methods

Recruitment

Participants were recruited from UK family support groups (the Smith-Magenis Foundation UK and Down's Syndrome Association) and an existing participant database at the Cerebra Centre for Neurodevelopmental Disorders. Inclusion criteria required confirmed diagnosis from a relevant professional (e.g. paediatrician or clinical geneticist).

Participants

Table 1 describes the sample demographics. Twenty two children with SMS and 21 with DS were recruited. One child with SMS could not be assessed due to challenging behaviour, one child with DS was excluded after validity checks indicated that an experimental manipulation was not upheld during testing[1] and for another child with DS the measure of ability could not be obtained. The final sample comprised 21 children with SMS and 19 with DS. Twenty children with SMS had a chromosome 17p11.2 deletion, one had a gene RAI1mutation. All children with DS had trisomy 21.

Participants were comparable for chronological age, gender and estimates of adaptive functioning (derived from the Vineland Adaptive Behavior Scales II), as shown in table 1.

[Insert table 1 about here]

[1] The participant's caregiver spoke to the child throughout each of the low attention conditions, exceeding the 30% cut off implemented to check the validity of experimental manipulations of levels of attention provided by the adults.

Measures

Informant report measures

A demographic questionnaire provided information on diagnosis, gender and age. Ability was assessed using the Vineland Adaptive Behavior Scales II – Interview edition, Survey form (Sparrow, Balla, & Cicchetti, 1984). Four domains measure adaptive behaviour (subdomains in parentheses) - Communication (receptive, expressive, written), Daily-living skills (personal, domestic, community), Socialisation (interpersonal relationships, play and leisure time, coping skills) and Motor skills (fine, gross). It has robust test-retest and inter-rater reliability and good internal consistency (Sparrow et al., 1984). Age equivalent scores from the subdomains of the Communication, Daily Living Skills and Socialisation domains are reported.

Direct observation

Level of attention was manipulated using a protocol based on the LabTab Parental Separation task (Goldsmith & Rothbart, 1993). In the original task mother and child play with toys (high attention: high levels of attention provided to the child), the mother then tells the child to stay and play and that she will return. The mother leaves (low attention: low levels of attention provided to the child), returning after 30 seconds. For this study the task was extended to include an episode of separation from an unfamiliar adult, to examine the effect of familiarity.

A further extension to the paradigm was the inclusion of a second ‘unresponsive’ adult. This adult remained in view of the child throughout high and low attention conditions but was instructed not to interact (ensuring that attention would remain reduced in low attention conditions). This enabled examination of whether children would attempt to utilise an

alternative source of attention, potentially providing insight into the phenomenology of 'attention seeking' behaviour in SMS, where excessive focus on one particular person is reported. Thus, each adult can either be interactive or unresponsive. To provide opportunity to approach the unresponsive adult, and optimise likelihood of more able children responding, time left playing alone was extended.

Conditions were five minutes long, with six conditions in total (see table 2), run consecutively. Testing was repeated three times to account for variation in behaviour. This repetition is recommended in the LabTab manual. Breaks were provided between each of the repetitions.

[Insert table 2 about here]

If the child approached the interactive adult during low attention conditions or the unresponsive adult in any condition, the adult indicated they were busy and instructed the child to play alone. The order of which adult played with the child first was counterbalanced.

Testing materials

Mothers were asked to provide toys that would be engaging and encourage social interaction. Observations were recorded on a Sony TRV-48E handheld camera.

Procedure

Testing and ethical considerations

Ethical review was obtained from the ethics committee of [withheld for blind review]. Informed consent for participation was gained from caregivers. Participants were observed at home, except one participant who was observed in school. Testing episodes were filmed, with the camera operator instructed to be inconspicuous but provide procedural prompts.

Real time coding

Chevallier et al. (2012) suggest that social motivation has three behavioural manifestations: orienting to social stimuli, seeking and liking social stimuli, and strategies employed to maintain interaction. In this study we coded child behaviours including adult directed looking (orienting), physical initiation of interaction (seeking) and affective responses suggesting level of enjoyment (liking) of social episodes. Maintaining behaviours were not coded as these are more complex (e.g. ingratiating).

Behaviour was coded using ObsWin 32 software (Martin, Oliver, & Hall, 2001), a software package recording frequency and duration of defined behaviours. Inter-rater reliability of behaviour definitions was calculated for 15% of the sample. Kappa values based on 5s intervals for all variables coded ranged from 0.8-1.0 (mean = .86), indicating good reliability (Fleiss, 1981). Operational definitions and Kappa values for all variables coded are shown in Online Resource 1. Values derived for each child behavioural variable reflect percentage of the observation the child engaged in a behaviour. Composite variables represent the sums of these percentages.

Several child behaviour variables were combined, creating a composite variable labelled physical initiation. This consisted of the sum of child approaching adult (kappa for approaching mother = .85, kappa for approaching researcher = .95), touching adult (kappa for

touching mother = .87, kappa for touching researcher = .87) and reaching to adult (kappa for reaching to mother = .86, kappa for reaching to researcher = .97). Child looking to adult was also coded (kappa for looking to mother = .81, kappa for looking to researcher = .8). Subsequently, two broad composite outcome variables were derived. 'Initiating' (sum of physical initiation and looking) assessed orienting and seeking behaviours. 'Enjoyment' (child positive affect; kappa = .71) evaluated children's liking of social situations. Integrity of experimental manipulations was evaluated by coding adult and child behaviours indicative of divergence from the protocol. Challenging behaviours (self-injury, aggression, property destruction) and negative affect were coded together with adult vocalisation and adult demands (e.g. to return if the child leaves the observation setting to access a tangible item).

Conditions were excluded if the following occurred for over 30% of the condition: challenging behaviour/negative affect carried over from a previous condition, challenging behaviour or negative affect unrelated to the experimental manipulation (e.g. associated with denials of a tangible item) or off protocol adult verbalising to the child in low attention conditions.

Data analysis

Data from the three repeats of each condition were averaged. No significant differences were found in behaviours in the '*adult plays*' conditions and the '*adult returns*' conditions, therefore the mean was calculated, resulting in one *high attention* condition for each adult.

As data were non-normally distributed non-parametric alternatives to ANOVA were employed to examine whether children with SMS and DS differed in their response to changes in levels of attention and familiarity (analogous to an interaction in ANOVA).

'Difference scores' (for attention: *low attention* behaviours subtracted from *high attention* behaviours, for familiarity: behaviours shown during interactions with the *unfamiliar adult* minus those with the *mother*) were calculated and compared between syndromes. Hypothesised differential effects of attention and familiarity depending on syndrome group would be demonstrated by significant between group differences in the magnitudes of 'difference scores' (analogous to a significant interaction between syndrome and condition).

Where a significant overall between syndrome effect of condition was found, two-tailed post hoc analyses of the original data established the underlying pattern of differences, using a more conservative alpha of ≤ 0.01. Comparisons were carried out separately for interactive and unresponsive adults.

Comparisons of enjoyment of interaction with adults of differing familiarity were conducted for *high attention* conditions only, as in *low attention* it is unclear who engendered this response.

Means and standard errors are displayed in figures as they most clearly illustrate patterns of results.

Results

Effect of decreased attention

The first hypothesis (hypothesis 1) was that initiation of interaction when attention is unavailable compared to available would be greater in SMS than DS. The magnitude of

differences between initiations[2] shown in high and low attention conditions were therefore compared across groups to explore between syndrome differences in responses to differing levels of attention. Table 3 shows the average difference between initiations made in low attention and high attention conditions ('High-low difference' rows).

[Insert table 3 about here]

A differential effect of attention dependent on syndrome group (indicating an interaction between attention and syndrome) was found for initiations towards the interactive adult for both *mothers* and the *unfamiliar adult* with a greater difference found between high and low attention conditions in DS than in SMS for initiations towards both adults. Both of these differences were associated with a medium effect size (see table 3).

Post hoc analyses demonstrated that in both *high* and *low attention* conditions there were no significant differences between SMS and DS in children's initiations towards their *mother* or an *unfamiliar adult* (shown in figure 1, left and right panels and in table 3 for between groups comparisons). Within-syndrome analyses showed that, as expected, both syndrome groups made significantly fewer initiations during *low attention* than in *high attention* toward both their *mother* (SMS, $Z = -3.702$, $p < .001$, $r = .57$; DS, $Z = -3.823$, $p < .001$, $r = .62$) and the *unfamiliar adult* (SMS, $Z = -4.015$, $p < .001$, $r = .62$; DS, $Z = -3.823$, $p < .001$, $r = .62$). These differences were associated with a large effect size.

These analyses indicate that while both syndromes showed reduced initiations towards the interactive adult in *low attention*, the magnitude of this reduction was less for SMS than DS. A smaller difference between high and low attention in SMS is consistent with hypothesis 1 which stated that, relative to when attention was available, initiations of interaction when

[2] See text in method section (real time coding procedure) for description of the derivation of the composite index of initiation

attention was unavailable would be greater in SMS than in DS. However, lack of significant between syndrome post hoc tests indicates that this effect depends on the *relative* differences between high and low attention.

Effect sizes of post hoc analyses examining behaviour in *high attention* and *low attention* conditions provide insight into this. For between syndrome comparisons involving the *unfamiliar adult* there was a large difference between effect sizes in the *high attention* condition compared to the *low attention* condition (.26 compared to .002). For initiations towards the *mother* this difference was much smaller (.18 compared to .09). For within syndrome comparisons it is only for interactions with the *mother* in SMS where effect sizes implicate a smaller difference between *high* and *low attention* conditions (.57 for the *mother* compared to .62 with the *unfamiliar adult* in SMS and .62 for both the *mother* and the *unfamiliar adult* in DS). This suggests that the significant overall effects found for the *unfamiliar adult* may be driven by between syndrome differences in initiations in *high attention.* The significant overall effects found for the *mother* may be driven by within syndrome differences in SMS between *high attention* and *low attention* conditions.

[Insert figure 1 about here]

No differential effect of reduced attention dependent on syndrome group was found for initiations directed towards the unresponsive adult. Data for these analyses are shown in the table in Online Resource 2.

Hypothesis 1 was therefore partially supported as a differential effect of reduced attention dependent on syndrome group was only found for initiations towards the interactive adult.

No differential effect of attention dependent on syndrome group was found for enjoyment, failing to support the hypothesis that enjoyment when attention was available (compared to

unavailable) would be greater in SMS than DS (hypothesis 2). Data for these analyses are shown in the table in Online Resource 3.

.

Effect of differing familiarity

The first hypothesis relating to familiarity (hypothesis 3) was that initiation of interaction with *mothers* (compared to an *unfamiliar adult*) would be greater in SMS than DS. The magnitude of differences between initiation behaviours directed towards each adult were therefore compared across groups to explore between syndrome differences in responses to differing levels of familiarity.

No differential effect of familiarity dependent on syndrome group was found for initiations directed towards the interactive adult, failing to support hypothesis 3 in this respect. Data for these analyses are shown in the table in Online Resource 4.

However, for the unresponsive adult, initiations directed towards *mothers* compared to the *unfamiliar adult* differed between syndromes, indicating an interaction between familiarity of the unresponsive adult and syndrome. Table 4 shows the average difference between initiations towards unresponsive *mothers* and *unfamiliar adults* ('Mother-unfamiliar adult difference' rows).

[Insert table 4 about here]

Relative to children with DS, children with SMS showed a greater difference between initiations towards their unresponsive *mother* compared to the unresponsive *unfamiliar adult*

in both *high attention* and *low attention* conditions. These differences were associated with a medium effect size (see table 4).

For behaviour in *high attention* conditions, post hoc analyses, illustrated in figure 2 (left panel) and table 4 for between groups comparisons, showed no significant differences between SMS and DS in children's initiations toward either their *mother* or the *unfamiliar adult*. Within-syndrome analyses found no significant difference between initiations toward *mothers* compared to *unfamiliar adults* in DS ($Z = -.236$, $p = .813$, $r = .04$). However, children with SMS made fewer initiations towards the *unfamiliar adult* than towards their *mothers* ($Z = -2.945$, $p = .003$, $r = .45$), a difference associated with a medium effect size.

[Insert figure 2 about here]

When experiencing *low attention* conditions (shown in figure 2, right panel and table 4 for between group comparisons), post hoc analyses found similar levels of initiations towards children's *mother* in SMS and DS, however significantly fewer initiations were made toward the *unfamiliar adult* by children with SMS, a difference associated with a large effect size. Within-syndrome analyses found no significant difference between initiations toward *mothers* compared to *unfamiliar adults* in DS ($Z = -.345$, $p = .73$, $r = .06$), whereas children with SMS made more initiations towards their *mother* than the *unfamiliar adult* ($Z = -2.982$, $p = .003$, $r = .46$), a difference associated with a medium effect size. These analyses show that, compared to children with DS, children with SMS made fewer initiations towards the *unfamiliar adult* as an alternative source of interaction than they did towards their *mothers*, even when experiencing low levels of attention. This preference is consistent with hypothesis 3, that initiation of interaction with *mothers* (compared to an *unfamiliar adult*) will be greater in SMS than DS. Overall hypothesis 3 is therefore partially supported as initiations of

interaction with *mothers* (compared to an *unfamiliar adult*) were greater in SMS than DS when the adult involved was the unresponsive adult but not when it was the interactive adult.

The final hypothesis (hypothesis 4), that positive affect during high attention conditions involving interactions with *mothers* (compared to an *unfamiliar adult*) would be greater in SMS than DS was not supported. No differential effect of familiarity dependent on syndrome group was found for positive affect shown during *high attention* conditions. Data for these analyses are shown in the table in Online Resource 5.

.

Discussion

We investigated motivation as a feature of social functioning which may differentiate neurodevelopmental disorders. Social motivation in children with SMS (characterised by heightened social motivation associated with negative behavioural outcomes), was directly observed and contrasted to DS (characterised by strong social motivation but fewer associated behavioural problems). Robust experimental methodology was used to systematically examine components of motivation. Extension of paradigms used in past research enabled more nuanced consideration of motivation in situations which reflect the complexity of 'real world' social situations (responses to reduced availability of social interaction and behaviour where attention is available from multiple sources).

Between syndrome differences in child responses to manipulation of social variables were found, supporting two out of four hypotheses stated. In line with hypothesis 1, initiation of interaction towards the interactive adult when attention was unavailable (compared to available) was greater in SMS than DS (although this pattern was not also found for initiations towards the unresponsive adult). Consistent with hypothesis 3, preference for

initiation of interaction with mothers (compared to an unfamiliar adult) was greater in SMS than DS. Hypotheses 2 and 4 regarding the effect of experimental manipulations on positive affect shown by children were not supported however. Notably, divergence in profiles of social behaviour between syndromes was not characterised by overall differences reflecting undifferentiated elevated social motivation in SMS compared to DS. Instead it was characterised by differences related to familiarity and available attention levels suggesting differences along nuanced dimensions to social motivation.

As children with SMS reportedly 'seek' attention (Haas-Givler, 1994) it was anticipated that creating a low attention situation (by an interactive adult leaving) would result in efforts to seek interaction by trying to initiate interactions with the adult who left, and potentially by approaching a remaining, unresponsive adult. Findings supported this to an extent. When attention was unavailable children with SMS did not show reduced approaches to the interactive adults to the same extent as children with DS (relative to their level of approaches in high attention conditions). This interpretation should be qualified by consideration of familiarity. For behaviours towards the child's mother, the smaller difference between high and low attention conditions in SMS more clearly potentially involves a relatively high rate of initiations in low attention when contrasted to DS. However, during situations involving the unfamiliar adult, the smaller difference seems to be dependent on low levels of initiations by children with SMS in high attention situations.

Lack of significant post hoc between syndrome comparisons suggests no absolute between group differences in initiations. Objectively therefore social motivation in children with SMS was not characterised by generalised increased motivation to interact with adults relative to children with DS, as found in some studies comparing 'hypersociable' Williams Syndrome with DS (Doyle et al., 2004; Jones et al., 2000). In fact average scores indicate that children

with SMS made *fewer* initiations than children with DS when experiencing high levels of attention. Only when level of available attention was lower did this pattern reverse in some instances. In SMS then, manifestation of heightened social motivation reported by caregivers may be dependent on specific variables, including *changes* in the level of attention available, consistent with reports of sensitivity to reduced availability of attention (Taylor & Oliver, 2008).

Interestingly, relatively increased motivation to engage with the interactive adult in low attention conditions was not reflected by increased utilisation of alternative sources of attention (the second, unresponsive adult). Relative to DS, lower levels of attention did not induce children with SMS to make more initiations towards the unresponsive adult. This finding may be understood in the context of existing reports of behaviour in individuals with SMS suggesting an excessive focus on one person (Moss et al., 2009), such that an alternative source of attention is unlikely to suffice if attention from a person who is currently the focus of their motivation becomes unavailable.

In addition to effects of level of attention, effects of familiarity of the adult were found. Both when experiencing high levels of attention from the interactive adult and when left alone in the room with the unresponsive adult, children with SMS made fewer initiations towards the unresponsive unfamiliar adult than towards their mother. This was not evident in DS. Indeed, when children were left to play alone with the unresponsive adult present, children with SMS initiated fewer interactions towards the unfamiliar adult than children with DS (with a mean close to zero), suggesting that they simply did not access the unfamiliar adult as an alternative source of attention when left alone by their mother. Overall, findings suggest a relatively strong preference in SMS for initiating interactions with mothers in preference to unfamiliar adults.

These nuanced between group differences in behaviour potentially provide insight into the issue of why strong social motivation is associated with more difficulties in SMS. Social behaviour of children with SMS may be less flexible than in DS, with less effective adaptation to different social situations implicated. Social initiations may persist when attention is no longer available and children may fail to utilise an alternative source of attention when a preferred adult is unavailable. In terms of the wider impact of this, attention-seeking directed towards a highly preferred familiar person, such as a caregiver, may be particularly demanding. In SMS this may contribute to the high reported stress levels of parents and carers (Hodapp, Fidler, & Smith, 1998). Such a relationship between targeted attention seeking and caregiver stress has been proposed in Angelman syndrome (Isles, 2011), where strong social motivation is also preferentially directed towards familiar adults (Mount et al., 2011) and caregivers have higher rates of anxiety than caregivers of those with other genetic syndromes (Griffith et al., 2011).

Given differential between syndrome effects of both attention and familiarity on initiation of interaction, it is notable that signs of enjoyment in children with SMS were not similarly dependent on these factors. This discrepancy contrasts with Angelman and Williams syndromes, where positive affect in response to social attention is characteristic (Jones et al., 2000; Oliver et al., 2007). Apparently children's motivation to interact was not manifested by elevated levels of 'typically' sociable behaviours such as positive affect ('liking' in Chevallier et al.'s model); instead, motivation was evidenced by increased orienting and seeking behaviours. This indicates that indices of social motivation may manifest differently across different neurodevelopmental disorders.

Lack of a mental age matched typically developing contrast group limits conclusions regarding the question of whether social motivation in SMS is objectively atypical (Hodapp

& Dykens, 2001). This clearly represents a limitation of this study. For example if children with DS are excessively sociable this would influence interpretation of results for SMS in this context. Inclusion of a typically developing contrast group which could address this issue is not without its difficulties however. Comparisons of children with SMS to typically developing children are themselves potentially limited by differences in chronological age likely to impact on social behaviour. Comparison of a typically developing 4 year old with an 11 year old with ID for example may be confounded by substantially different experiences of social situations (amount of experience gained over the lifetime by older children, how adults respond to older children with ID). The 'same but different' syndrome contrast approach employed in the current study circumvents this issue and instead answers a different question around how two syndromes reported to have high social motivation may differ in how this manifests (Hodapp & Dykens, 2001). It is crucial, however, to emphasise that the current study does not provide evidence that social motivation is atypical in SMS, only that it is different from DS.

Bearing this limitation in mind, the results of the current study provide novel empirical evidence that social motivation in SMS differs from another syndrome also associated with strong social motivation but where difficulties associated with this are less frequently described (DS). This difference is characterised by comparatively frequent social initiations in low attention conditions and stronger preference for familiar adults, responses which may account in part for why social motivation is reported to be more problematic in SMS than DS. More broadly, these findings suggest a need for consideration of specific *dimensions* of social motivation, as more nuanced features of the phenomenology of social motivation may not be captured by variations along a broad excessive-impaired dimension. This includes preferential motivation and divergence of indices of motivation, such as initiation of interaction versus enjoyment of social situations.

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards

References

Chen, R. M., Lupski, J. R., Greenberg, F., & Lewis, R. A. (1996). Ophthalmic manifestations of Smith-Magenis syndrome. *Ophthalmology, 103*(7), 1084-1091.

Chevallier, C., Kohls, G., Troiani, V., Brodkin, E. S., & Schultz, R. T. (2012). The social motivation theory of autism. *Trends in Cognitive Sciences, 16*(4), 231-239.

Cook, F., & Oliver, C. (2011). A review of defining and measuring sociability in children with intellectual disabilities. *Research in Developmental Disabilities, 32*(1), 11-24.

Davies, M., Udwin, O., & Howlin, P. (1998). Adults with Williams syndrome. Preliminary study of social, emotional and behavioural difficulties. *The British Journal of Psychiatry, 172*(3), 273-276.

Dawson, G., Meltzoff, A., Osterling, J., Rinaldi, J., & Brown, E. (1998). Children with autism fail to orient to naturally occurring social stimuli. *Journal of Autism and Developmental Disorders, 28*(6), 479-485. doi: 10.1023/a:1026043926488

Dykens, E. M., Finucane, B. M., & Gayley, C. (1997). Brief report: Cognitive and behavioral profiles in persons with Smith-Magenis syndrome. *Journal of Autism and Developmental Disorders, 27*(2), 203-211. doi: Doi 10.1023/A:1025800126086

Dykens, E. M., & Kasari, C. (1997). Maladaptive behavior in children with Prader-Willi syndrome, Down syndrome, and nonspecific mental retardation. *American Journal on Mental Retardation, 102*(3), 228-237.

Dykens, E. M., & Smith, A. C. M. (1998). Distinctiveness and correlates of maladaptive behaviour in children and adolescents with Smith-Magenis syndrome. *Journal of Intellectual Disability Research, 42*, 481-489. doi: DOI 10.1046/j.1365-2788.1998.4260481.x

Fleiss, J. L. (1981). *Statistical Methods for Rates and Proportions*. NewYork: John Wiley and Sons.

Goldsmith, H., & Rothbart, M. (1993). The laboratory temperament assessment battery (LAB-TAB). *University of Wisconsin*.

Gosch, A., & Pankau, R. (1997). Personality characteristics and behaviour problems in individuals of different ages with Williams syndrome. *Developmental Medicine & Child Neurology, 39*(8), 527-533.

Greenberg, F., Lewis, R. A., Potocki, L., Glaze, D., Parke, J., Killian, J., . . . Lupski, J. R. (1996). Multi-disciplinary clinical study of Smith-Magenis syndrome (deletion-17p11.2*).* *American Journal of Medical Genetics, 62*(3), 247-254. doi: Doi 10.1002/(Sici)1096-8628(19960329)62:3<247::Aid-Ajmg9>3.3.Co;2-9

Grelotti, D. J., Gauthier, I., & Schultz, R. T. (2002). Social interest and the development of cortical face specialization: What autism teaches us about face processing. *Developmental Psychobiology*, 40(3), 213-225. doi: 10.1002/dev.10028

Griffith, G. M., Hastings, R. P., Nash, S., & Hill, C. (2010). Using matched groups to explore child behavior problems and maternal well-being in children with Down syndrome and autism. *Journal of Autism and Developmental Disorders, 40*(5), 610-619.

Griffith, G. M., Hastings, R. P., Oliver, C., Howlin, P., Moss, J., Petty, J., & Tunnicliffe, P. (2011). Psychological well-being in parents of children with Angelman, Cornelia de Lange and Cri du Chat syndromes. *Journal of Intellectual Disability Research, 55*(4), 397-410. doi: 10.1111/j.1365-2788.2011.01386.x

Haas-Givler, B. (1994). Educational Implications and Behavioral Concerns of SMS—From the Teacher's Perspective. *Spectrum (Newsletter of PRISMS), 1*(2), 3-4.

Hodapp, R. M., & Dykens, E. M. (2001). Strengthening behavioral research on genetic mental retardation syndromes. *American Journal on Mental Retardation, 106*(1), 4-15.

Hodapp, R. M., Fidler, D., & Smith, A. (1998). Stress and coping in families of children with Smith-Magenis syndrome. *Journal of Intellectual Disability Research, 42*(5), 331-340.

Hodapp, R. M., Ly, T. M., Fidler, D. J., & Ricci, L. A. (2001). Less stress, more rewarding: Parenting children with Down syndrome. *Parenting: Science and Practice, 1*(4), 317-337.

Jones, W., Bellugi, U., Lai, Z., Chiles, M., Reilly, J., Lincoln, A., & Adolphs, R. (2000). II. Hypersociability in Williams syndrome. *Journal of Cognitive Neuroscience, 12*(1), 30-46.

Kasari, C., & Freeman, S. F. N. (2001). Task-related social behavior in children with Down syndrome. *American Journal on Mental Retardation, 106*(3), 253-264. doi: 10.1352/0895-8017(2001)106<0253:trsbic>2.0.co;2

Kasari, C., Mundy, P., Yirmiya, N., & Sigman, M. (1990). Affect and attention in children with Down syndrome. *American Journal on Mental Retardation, 95*(1), 55-67.

Langthorne, P., & McGill, P. (2012). An indirect examination of the function of problem behavior associated with fragile X syndrome and Smith-Magenis syndrome. *Journal of Autism and Developmental Disorders, 42*(2), 201-209. doi: DOI 10.1007/s10803-011-1229-6

Martin, G. E., Klusek, J., Estigarribia, B., & Roberts, J. E. (2009). Language characteristics of individuals with Down syndrome. *Topics in Language Disorders, 29*(2), 112.

Martin, N., Oliver, C., & Hall, S. (2001). Obswin: Software for the collection and analysis of observational data. *Birmingham, UK: University of Birmingham*.

Melyn, M. A., & White, D. T. (1973). Mental and developmental milestones of noninstitutionalized Down's syndrome children. *Pediatrics, 52*(4), 542-545.

Moss, J., & Howlin, P. (2009). Autism spectrum disorders in genetic syndromes: implications for diagnosis, intervention and understanding the wider autism spectrum disorder population. *Journal of Intellectual Disability Research, 53*(10), 852-873. doi: 10.1111/j.1365-2788.2009.01197.x

Moss, J., Oliver, C., Arron, K., Burbidge, C., & Berg, K. (2009). The prevalence and phenomenology of repetitive behavior in genetic syndromes. *Journal of Autism and Developmental Disorders, 39*(4), 572-588. doi: DOI 10.1007/s10803-008-0655-6

Mount, R., Oliver, C., Berg, K., & Horsler, K. (2011). Effects of adult familiarity on social behaviours in Angelman syndrome. *Journal of Intellectual Disability Research, 55*(3), 339-350.

Mundy, P., Sigman, M., Kasari, C., & Yirmiya, N. (1988). Nonverbal communication skills in Down syndrome children. *Child Development, 59*(1), 235-249.

Oliver, C., Demetriades, L., & Hall, S. (2002). Effects of environmental events on smiling and laughing behavior in Angelman syndrome. *American Journal on Mental Retardation, 107*(3), 194-200.

Oliver, C., Horsler, K., Berg, K., Bellamy, G., Dick, K., & Griffiths, E. (2007). Genomic imprinting and the expression of affect in Angelman syndrome: what's in the smile? *Journal of Child Psychology and Psychiatry, 48*(6), 571-579.

Porter, M. A., Coltheart, M., & Langdon, R. (2007). The neuropsychological basis of hypersociability in Williams and Down syndrome. *Neuropsychologia, 45*(12), 2839-2849.

Slager, R. E., Lynn, T., Newton, T. L., Vlangos, C. N., Finucane, B., & Elsea, S. H. (2003). Mutations in RAI1 associated with Smith-Magenis syndrome. *Nature Genetics, 33*(4), 466-468. doi: Doi 10.1038/Ng1126

Sloneem, J., Oliver, C., Udwin, O., & Woodcock, K. A. (2011). Prevalence, phenomenology, aetiology and predictors of challenging behaviour in Smith-Magenis syndrome. *Journal of Intellectual Disability Research, 55*(2), 138-151.

Smith, A. C., McGavran, L., Robinson, J., Waldstein, G., Macfarlane, J., Zonona, J., . . . Magenis, E. (1986). Interstitial deletion of (17)(p11.2p11.2) in nine patients. *American Journal of Medical Genetics, 24*(3), 393-414. doi: 10.1002/ajmg.1320240303

Smith, A. C. M., Dykens, E., & Greenberg, F. (1998). Behavioral phenotype of Smith-Magenis syndrome (del 17p11.2). *American Journal of Medical Genetics, 81*(2), 179-185. doi: Doi 10.1002/(Sici)1096-8628(19980328)81:2<179::Aid-Ajmg10>3.0.Co;2-E

Sparrow, S. S., Balla, D. A., & Cicchetti, D. V. (1984). *Vineland adaptive behavior scales: Interview edition, survey form manual*: American Guidance Service Circle Pines, MN.

Taylor, L., & Oliver, C. (2008). The behavioural phenotype of Smith-Magenis syndrome: evidence for a gene-environment interaction. *Journal of Intellectual Disability Research, 52*(10), 830-841. doi: DOI 10.1111/j.1365-2788.2008.01066.x

Wilde, L., Silva, D., & Oliver, C. (2013). The nature of social preference and interactions in Smith–Magenis syndrome. *Research in Developmental Disabilities, 34*(12), 4355-4365.

Wing, L., & Gould, J. (1979). Severe impairments of social interaction and associated abnormalities in children: Epidemiology and classification. *Journal of Autism and Developmental Disorders, 9*(1), 11-29.

Figure Captions

Figure 1. Post hoc analysis of initiations made towards interactive mothers (left panel) and unfamiliar adults (right panel) in high and low attention conditions (** $p \leq .001$). Solid line represents within groups comparisons.

Figure 2. Post hoc analyses of initiations to the unresponsive adults made in *high attention* (left panel) and *low attention* (right panel) conditions * $p \leq .01$, ** $p \leq .001$). Solid line represents within groups comparisons; dashed line represents between groups comparisons.

Figure 1 top

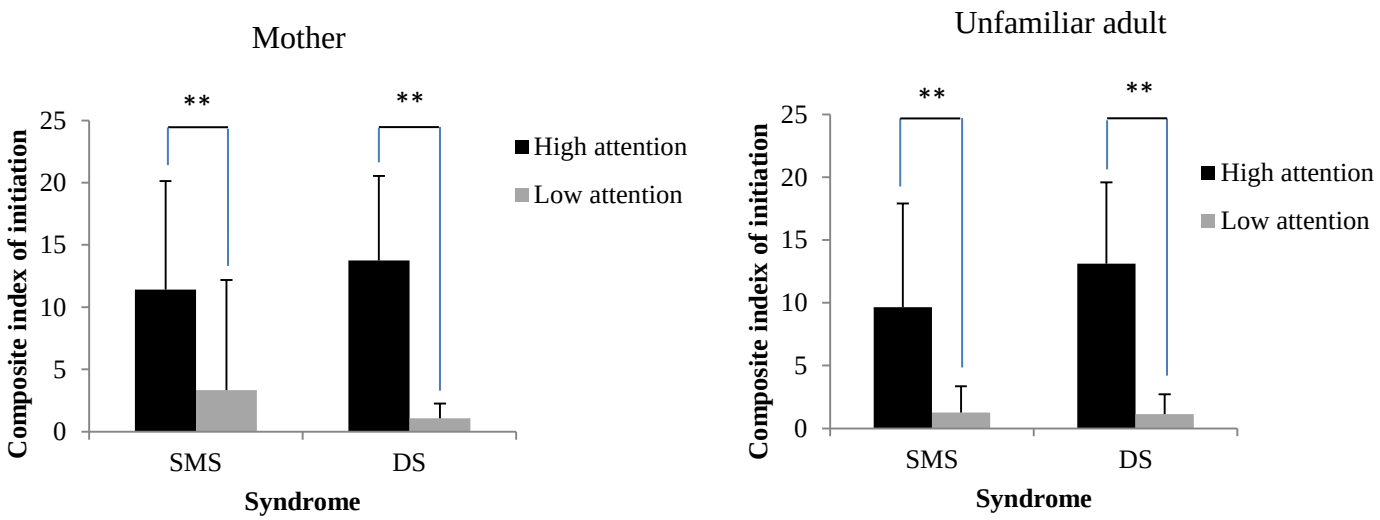
Mother
Unfamiliar adult
Composite index of initiation
Composite indeix of initiation
25
20
15
10
5
0
**
**
SMS
DS
Syndrome
High attention
Low attention

Figure 2 top

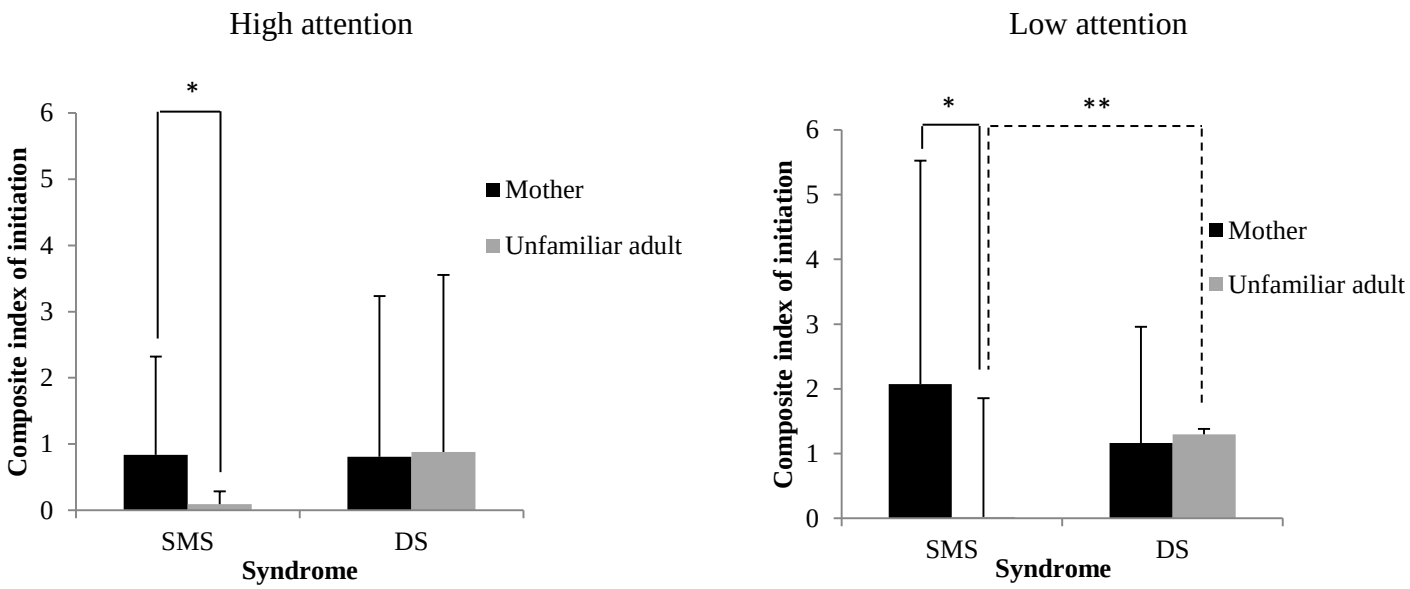
High attention
Low attention
Composite index of initiation
Syndrome
SMS
DS
Mother
Unfamiliar adult
*
**
0
1
2
3
4
5
6

Table 1. Participant demographic characteristics and age equivalent (AE) scores obtained on Communication, Daily Living Skills and Socialisation domains of the Vineland Adaptive Behavior Scales II (VABS), for each syndrome group

			Smith-Magenis syndrome	Down syndrome	Between groups comparison
N			21	19	
Age		Median	76.71	84.96	U = 184, p = .675
(Months)		*(Range)*	*(30.92-189.80)*	*(38.80-157.83)*	
Gender		Number male	12	10	χ^2 = .082, p = .512
		(%)	*(57.14)*	*(52.63)*	
VABS domains and subdomains					
Communication	Receptive[1]	Mean AE	27.52	34.95	t(38) = -1.72, p = .093
		(SD)	*(11.61)*	*(15.52)*	
	Expressive[1]	Mean AE	31	33.84	t(38) = -.45, p = .654
		(SD)	*(24.08)*	*(13.67)*	
	Written	Mean AE	54.57	*59.79*	t(38) = -.59, p = .556
		(SD)	*(33.31)*	*(19.86)*	
Daily Living Skills	Personal	Mean AE	35.67	38.47	t(38) = -.51, p = .615
		(SD)	*(21.07)*	*(13.37)*	
	Community	Mean AE	49.43	47.00	t(38) = .25, p = .801
		(SD)	*(36.38)*	*(21.34)*	
	Domestic[1]	Mean AE	52.10	52.68	t(38) = -.05, p = .962
		(SD)	*(44.27)*	*(30.96)*	
Socialisation	Interpersonal				

relationships[1]	Mean AE	29.9	35.74	t(38) = -.78, *p* = .440
	(SD)	*(29.39)*	*(14.76)*	
Play and leisure time[2]	Mean AE	38.45	50.11	t(38) = -1.65, *p* = .107
	(SD)	*(23.56)*	*(20.27)*	
Coping skills	Mean AE	32.52	33.05	t(38) = -.09, *p* = .932
	(SD)	*(24.55)*	*(13.22)*	

[1] Data for these subdomains were non-normally distributed (Shapiro-Wilk tests p <.05). However, Mann-Whitney U tests (alpha = .01 because of multiple analyses), also found no significant differences: Receptive U = 147, p = .154; Expressive, U = 163.5, p = .328; Domestic, U = 181.5, p = .625; Interpersonal relationships, U = 121, p = .033

[2]Data missing for one participant with SMS

Table 2. Experimental conditions employed in observations

Overall condition	Interactive adult	Level of attention	Unresponsive adult	Adult behaviour
Mother plays	Mother	High attention	Unfamiliar adult	Mother engages child in play (unfamiliar adult present but ignores child)
Mother leaves	Mother	Low attention	Unfamiliar adult	Mother leaves room (unfamiliar adult remains in room ignoring child)
Mother returns	Mother	High attention	Unfamiliar adult	Mother reengages child in play (unfamiliar adult still present but ignoring child)
Unfamiliar adult plays	Unfamiliar adult	High attention	Mother	Unfamiliar adult engages child in play (Mother present but ignores child)
Unfamiliar adult leaves	Unfamiliar adult	Low attention	Mother	Unfamiliar adult leaves child (mother remains in room ignoring child)
Unfamiliar adult returns	Unfamiliar adult	High attention	Mother	Unfamiliar adult reengages child in play (mother still present but ignoring child)

Table 3. Initiations towards interactive adults in high and low attention conditions, the difference between these conditions (medians and range) and inferential analysis of between group differences

Interactive adult	Level of attention		Smith-Magenis syndrome	Down syndrome	Significance of between group differences
Mother	High				
		Median	8.98	11.67	U = 157, p = .25, r = .18
		Range	*(.56-32.78)*	*(3.33-39.95)*	
	Low				
		Median	1.11	0.93	U = 177.5, p = .549, r = .09
		Range	*(.00 -27.04)*	*(.00-4.63)*	
	High-low difference				
		Median	6.95	10.72	U = 125, *p* = .044, r = .32
		Range	*(-6.19 -30.01)*	*(3.33-37.73)*	
Unfamiliar adult	High				
		Median	6.39	13.43	U = 138, p = .096, r = .26
		Range	*(1.39-30.81)*	*(4.17-29.72)*	
	Low				
		Median	0.37	0.37	U = 199, p = .989, r = .002
		Range	*(.00-9.71)*	*(.00-5.38)*	
	High-low difference				
		Median	6.39	12.87	U = 126.5, *p* =.048, r = .31
		Range	*(.84 – 29.32)*	*(3.8-29.35)*	

Table 4. Initiations made towards unresponsive mothers and unfamiliar adults, the difference between these conditions (medians and range) and inferential analysis of between group differences

Level of attention	Unresponsive adult		Smith-Magenis syndrome	Down syndrome	Significance of between group differences
High	Mother				
		Median	0.46	0.09	U = 166.5, p = .438, r = .15
		Range	*(.00-6.39)*	*(.00-10.65)*	
	Unfamiliar adult				
		Median	0	0	U = 164, p = .248, r = .18
		Range	*(.00-.56)*	*(.00-11.57)*	
	Mother-unfamiliar adult difference				
		Median	0.46	0	U = 107, *p* = .01, r = .41
		Range	*(-.46-6.39)*	*(-.76-1.39)*	
Low	Mother				
		Median	0.56	0.56	U = 186.5, p = .714, r = .06
		Range	*(.00-13.33)*	*(.00-6.85)*	
	Unfamiliar adult				
		Median	0	0.37	U = 89.5, p < .001, r = .58
		Range	*(.00-.37)*	*(.00-7.04)*	
	Mother-unfamiliar adult difference				
		Median	0.56	0	U = 108, *p* = .011, r = .40
		Range	*(-.09-13.33)*	*(-3.89-3.70)*	